PARAMYOCLONUS MULTIPLEX, WITH A REPORT OF A CASE

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PARAMYOCLONUS multiplex is a spasmodic affection of the muscular system of peculiar character, distribution, and course, dependent upon irritation of the nervous motor mechanisms.

The chief features of this disease are illustrated by the following case:

John D., aged 33, of Kingston, Canada, a grocer, of good family history, but of nervous temperament, was in his usual health until September 15th, 1886, when he strained his back and right shoulder by lifting a box, sixty pounds in weight, while in an awkward position. The pain, below the right shoulder blade, resulting from this strain was so severe that he went at once to his physician, Dr. H. J. Saunders, in whose office he had a peculiar attack. He began to cry and scream with pain and soon felt a choking sensation and was unable to get his breath; then followed convulsive movements of the body and legs, the latter being drawn up and thrust out forcibly. These lasted an hour, after which he was taken home. They returned at short intervals, for three days, being attended by pain in the back; and then they began to involve the muscles of the upper extremities and neck; and a few twitching movements of the face also occurred. The spasms were always very rapid, but were chiefly confined to muscles attached to the trunk, it being noticed from the first that the muscles of the forearms and hands and of the legs and feet did not take part in the spasms. The diaphragm was affected usually, so that dyspnœa and

exhaustion attended the attack, and it was occasionally followed by vomiting, when occurring after a meal. He had at first ten or twelve such attacks in twenty-four hours, each lasting nearly an hour. After two or three weeks, the attacks became less severe, the legs being less violently moved, and their duration decreased. But they had continued until May 2d, 1887, when I saw him, varying in severity and duration, but constantly diminishing, so that now he may have but three or four a day, though sometimes they are more frequent. He had several attacks in my presence, each lasting from one to five minutes, and leaving him much exhausted. The muscles first affected were those of the back and abdomen, a series of quick, alternate contractions of the dorsal muscles and of the recti abdominis, resulting in a rapid protrusion and retraction of the abdomen. As this became more rapid and forcible, the body and head were thrown backward and forward, without any spasm of the muscles of the neck, and in one attack which occurred while he was standing, similar complementary movements to preserve his balance produced alternate slight flexion and extension of the hips. In a much more severe attack which was observed when he was stripped, the muscles involved were first those of the back and abdomen already mentioned; then of the neck, so that the head was not only nodded, but was turned from side to side; then of the muscles of the upper arm, the pectorals, deltoid, biceps, and triceps, being all in action, and lastly, the muscles of the thigh, the quadriceps femoris, biceps, and semi-tendinosus and semi-membranosus, with the glutei acting with such force as to cause movements of both hip and knee joints. The contractions of these muscles were rapid, the rate rising to ninety per minute. The spasm in the arms was not severe enough to move the shoulder or elbow joints, but the muscles named were seen to contract and raise the skin. He said that the wrist and fingers, ankles and toes, had never participated in the spasm. He had noticed that formerly there was some movement on the forearms, probably of the supinator longus muscles, but this I did not see.

There was also, in a severe attack, a spasm of the diaphragm resulting in a long inspiration, accompanied by a sound, and while this was tonic, rapid movements of the intercostal and accessory respiratory muscles to supply the lack of inspiratory action of the diaphragm were made. Only at the very first has there been seen any spasm of the facial muscles, and for some months these have not been affected.

The spasms were so severe as to make me fear that he would fall while standing, as he said he had done several times, and would nearly throw him out of an arm chair when sitting. They come on suddenly, but are usually preceded by a peculiar sensation which ascends from the legs to the head and ceases as suddenly, leaving him in a state of considerable exhaustion, panting and perspiring, and looking badly as if about to faint. During the interval between the spasms an occasional fibrillary twitching in the muscles of the back and pectorals was observed, but not elsewhere. He could not stop the spasm voluntarily or in any way limit its course. Nor could he start it voluntarily. But any exposure of the skin to cold, any irritation of the skin by electricity, any tapping of the tendons at the knee or attempt to elicit ankle clonus was sufficient to start a spasm at once. He said it often came on after muscular effort, such as a long walk. It was also more likely to occur after mental excitement, and it usually came on when he began to talk about his condition or went to see a stranger. From the beginning until the present time, pressure on the right shoulder blade, where he had the pain, produced a spasm at once. It ceased during his sleep and never woke him by occurring at night. He thought that a drink of whiskey arrested it sometimes.

His motor power, sensory perception, and voluntary co-ordination were not in any way impaired. His skin and tendon reflexes were exaggerated, a marked ankle clonus being present on both sides. His muscles responded normally to both electrical currents. Mentally, he was perfectly clear, but somewhat excitable, and after

each attack in his exhausted condition, tears came to his eyes as he spoke of his trouble. He did not appear, however, to be of an hysterical temperament. Treatment had been somewhat successful, as he considered his condition much better than it had been six months ago, for he said that the attacks were less severe, shorter, and less frequent.

This spasmodic affection, limited to the muscles of the body and proximal portion of the limbs and only occasionally affecting the neck and face, has been termed by Friedreich,[1] who described it first as paramyoclonus multiplex. Since its first description, only seven cases have been reported, those of Lowenfeld,[2] Marie,[3] Silvestrini,[4] Bechterew,[5] Seeligmuller,[6] Homen,[7] and Remak.[8] My own case brings the total number up to nine.* It must be regarded as a distinct disease, since it is widely different from chorea, from hysterical spasms, from epileptiform convulsions, from convulsive tremor or tetanilla, and from tic convulsif.

Its causation is uncertain. In three cases fright, in one case severe hemorrhage which may have caused fright, in one case the shock of a cold bath, and in my own case an overstrain causing pain have preceded the development of the spasm. In the other three cases, no cause could be found. In two of the cases, a chronic spasmodic affection had been present for several years. It has been observed but once in a female, the remaining eight cases being males. The age of the individual has little to do with its development, for persons of all ages from ten to fifty-two have been affected. Its symptoms are quite characteristic. The spasms are bilateral and symmetrical. They are limited to certain muscles. In eight cases, the quadriceps femoris and flexors of the leg, and the so-called "upper arm group of muscles," the deltoid, biceps, and supinator longus, were affected. In seven cases, the muscles of the back were involved. In six cases, the muscles of the neck contracted. In five cases, the glutei. In

* The case of Silvestrini is so imperfectly described in abstracts as to be useless for comparison, and the original was not accessible to me.

four cases, the face and the diaphragm were involved. In no case have the muscles of the hands or forearms, of the feet or legs been affected. The usual limitation of the spasm to the body muscles with those of the thighs and arms is very noticeable.

The character of the spasm is also characteristic. It is a rapidly repeated clonic spasm occurring at intervals. In six cases, the rate of contraction has been counted. It has varied from 50 per minute to 180 per minute. In my own case, it was about 90. It is not a sudden, single irregular muscular contraction, like that of chorea, but appears to be always bilateral and to involve several muscles of a physiological group at once, thus resulting in a series of movements, any one of which can be voluntarily made. In several cases, a tonic contraction has occurred in one or more of the muscles affected, before or during the clonic seizure. In my own case, the spasm of the diaphragm was tonic for one-eighth to one-quarter of a minute during each attack, and in the early attacks, the spasms of the back were tonic for some seconds. The clonic contractions continue, when once set up, for a varying time, from half a minute to ten minutes, and are succeeded by a complete interval of freedom from spasm. In my own case, this interval had varied from half an hour to about one week. And the fact that the free quiet intervals were getting longer had encouraged him to hope for a recovery. During the spasm itself, the resulting movements were of a very violent nature. The head was thrown about by the movements of the body, rendering the patient dizzy. The body was tossed about in the chair, so that there seemed to be danger of his being thrown out upon the floor. If the spasm occurred while he was walking, he was quite liable to be thrown down, and had hurt himself several times. But this violence is not always present—for in two cases, the spasms were never severe enough to cause a movement of the joints, and were only observed when the patient was stripped—being then of the nature of a fascicular twitching. In my own case, such a fascicular muscular twitch-

ing was occasionally seen during the intervals in the muscles of the back and the pectorals.

In the majority of the cases, any tapping of the tendons or any irritation of the skin was sufficient to produce a spasm. This seems to be an important point, for I am not aware that it has been observed in hysterical or choreic spasms. It is true that, in hysterical cases, certain zones or areas can occasionally be found on the body, irritation of which may cause or may arrest the attack. But in this condition the spasm is produced by irritation anywhere on the skin—or by tapping the tendons at the knee and ankle—and was not associated with disturbances of sensation, which are characteristic of hysterical zones. The knee jerk has been increased in four cases, was less in one case, and was not tested in the remainder. The skin reflexes were also increased in four cases, and are mentioned as normal in but one of the remainder. Mental excitement seems to have predisposed to the onset of the spasm in three cases. Had the disease been hysterical in nature, this would probably have been observed in a larger proportion. Voluntary effort stopped the spasm in four cases and made it worse in three cases. Had the disease been hysterical, volition would probably not have influenced it favorably in the majority of cases. The spasm has ceased during sleep in four cases, but has continued in one case.

In none of the cases have consciousness, motion, sensation, co-ordination, or electric excitability been in any way affected—an important negative fact, since it proves at once that the condition is a functional neurosis, and makes it very unlikely that it is of an epileptic or an hysterical nature. In one case, which died of phthisis, a careful examination by Prof. Schultze, of Heidelberg, failed to reveal any lesion of the nervous system.

It is evident from this review of the symptomatology that the characteristics of the disease are quite distinct; that it can be differentiated from chorea, from hysteria, and from epilepsy. Is there any disease known which it at all simulates? In tic convulsif, we have an affection of the

face consisting of spasmodic contractions of irregular intensity and frequency, often attended by intervals of freedom. The resemblance to paramyoclonus multiplex is more than superficial, and has been noticed by several writers. But all seem to agree that in tic convulsif the face is usually chiefly, if not exclusively, affected; that the contractions are often single and unilateral, are liable to occur during voluntary motion; that their intensity is not varied, but is quite uniform; that the spasm is not produced or increased by external influences; and that it is always a co-ordinated volitional motion which is produced. Guinon,* it is true, has described a *maladie des tics convulsifs*, in which title he wishes to include those spasmodic affections described by various authors as jumping, coprolalia, myriachit. But here again there is a wide difference from paramyoclonus multiplex. It seems, therefore, as though the disease must be regarded as distinct from tic convulsif, and as having a character of its own.

The question has arisen whether it is identical with an affection described by Hammond in 1867 as convulsive tremor. This was suggested by Dr. C. L. Dana, who saw my case and identified it as paramyoclonus multiplex. I have carefully studied the various cases described under this head by Hammond in his original article[9] and in the last edition of his work on "Nervous Diseases,"[10] and in a recent report of a case.[11] These cases are by no means uniform, and do not seem to me to belong to a single class of disease. There are but three which in any way resemble the disease under discussion. In two of these (cases V. and VI.), there was no limitation of the spasms to the the muscles affected in paramyoclonus—the arms and legs being affected as well as other muscles; there were motor and sensory symptoms, which were well marked, in addition to the spasms; and there were cerebral symptoms of such intensity as to lead the author originally to ascribe the disease to the cerebellum, a position which, however, he has retracted at present. It seems to me, therefore, that Hammond's original cases were either not paramyo-

* Rev. de Méd., 1886, January.

clonus multiplex or, if they were, no one from his description of them could arrive at the characteristic features of this disease. It is, therefore, necessary—much as we might prefer to claim for our own country the original observations—to admit that Friedreich was the first to describe the affection, and to adopt the name which he chose.

A case reported by Hammond only last year,[11] under the title of convulsive tremor, does, however, seem to conform to the picture of this disease. "A female, aged 22, was affected by an involuntary contraction of the gluteal and spinal muscles, by which the body was violently raised from the recumbent or sitting posture and, at the same time, drawn violently backward, and on the completion of this movement, the body, with equal violence and suddenness, was bent forward by the contraction of the abdominal and pelvic muscles, and then a period of rest of variable duration, sometimes being only a few seconds, and others several minutes, intervened. There were no other disturbances of mind or body, and no hysterical manifestations. Attacks were increased by mental excitement, and at a minimum when the mind was calm. Muscular activity did not influence them. They were absent during sleep." They could be controlled by the will only for a short time, the effort producing a feeling of nervousness. No cause was found for the condition which, after resisting bromides, yielded to hypodermatic injections of arsenic.

Here the picture is sufficiently distinct and conforms quite closely to that of paramyoclonus multiplex, while it differs widely from that of convulsive tremor, as described in the author's text-book. If this be accepted as a case of paramyoclonus multiplex, it brings the number of cases up to ten.

The only other cases of convulsive tremor on record are two, which were reported from Dr. Hammond's clinique by his assistant Dr. Brown;[12] but neither of them are cases of paramyoclonus multiplex. In one of them, a manifestly hysterical case, the spasm was limited to one arm. The other case is one of chorea electrica, first described

by Henoch. And as this disease might be mistaken for paramyoclonus multiplex by a superficial observer, it may be well to mention its characteristics. Henoch says[18] that in chorea electrica we have a combination of true choreic movements with clonic twitchings. The patients are quiet, and lightning-like twitches occur from time to time, perhaps every five minutes or more frequently, especially on the muscles of the neck and shoulders. There is also seen a twitching of individual muscles when the body is naked, not sufficient to move the limbs. These continue during sleep. The disease occurs between the ages of nine and fifteen, and is a manifestation of direct or reflex irritation of the nervous centres. To this description Dr. Brown's case corresponds quite closely, there being associated with a severe condition of chorea attacks of tonic, followed by clonic spasms, while in the interval fibrillary tremors were constantly observed. The spasms affected the head, hands, and feet, and implicated all the voluntary muscles, including the diaphragm and larynx, so that the patient would growl and bark. The tendon reflex was "slight," and there was "some anæsthesia," the location of which is not stated. The boy recovered under arsenical treatment.

It is evident that a number of different conditions have been brought together under the term convulsive tremor, and of the cases described I find but one in which the limitation of the spasm to certain muscles and the character of the attacks enables one to identify the disease as paramyoclonus multiplex.

But one other affection requires notice in connection with this disease. In a paper, read on Oct. 18th, 1886, before the Medical Society of the County of New York, Dr. Julius Althaus, of London, described an affection which he named tetanilla. He said that it was the same as paramyoclonus multiplex; that he had never published any observations on the subject up to that time, but that he had privately given the name tetanilla to the affection as more euphonious than that selected by Friedreich. He claimed to have seen a number of cases, and of these he alluded in a very cursory manner to five. Of these cases,

three were cases of unilateral spasm, and as one of the characteristics of paramyoclonus is the bilateral symmetrical nature of the convulsion, these cases may be ruled out at once. One of the remaining two cases is not sufficiently detailed to warrant any conclusion as to its nature. The last case (the first in his paper) cannot be considered one of the disease now under discussion. For Dr. Althaus says that all the muscles of the body were affected either simultaneously or successively, and the spasms commonly began in the muscles of the thumbs, each contraction lasting five or six seconds, and being succeeded by six or seven more contractions. Here there is no limitation of the spasms to the trunk muscles, no record of a series of rapid clonic spasms succeeded by a period of repose, no evidence of the presence of fibrillary tremors in the interval. It is evident that Dr. Althaus is mistaken in supposing that his cases were paramyoclonus multiplex. And the remark made by Dr. Hammond during the discussion, that he had not seen any such cases, leads to the conclusion that they were probably not of the nature of convulsive tremor.

It is evident from this review that the characteristic features of paramyoclonus multiplex have not been generally appreciated, and hence it has been mistaken for other affections, and diseases widely different from it, and only resembling it in respect to the common symptom of clonic spasms, have been classed with it. It may perhaps seem impossible from ten cases to form a picture of the disease, but doubtless these cases will be added to and the features of the affection more carefully outlined when its chief characteristics are more definitely grasped.

These characteristics may be summed up as follows: Paramyoclonus multiplex is a spasmodic affection of the muscular system, occurring bilaterally in symmetrically situated muscles attached at one or both ends to the trunk, and in muscles whose function is associated with these, consisting of a series of violent clonic spasms of considerable rapidity and severity, occurring only at intervals; and associated with fascicular tremors of the affected

muscles, persisting during the interval between the spasms. It occurs after some mental or physical strain, and is not accompanied by any disturbance of sensory or motor functions, excepting by an increase of the superficial and deep reflexes. It can be excited by irritation of the skin or tendons.

In regard to the prognosis, it may be said that this is favorable. The majority of the cases have recovered quite rapidly under treatment. In two cases, however, relapses have occurred.

The treatment which has been of most service has been the application of strong galvanic currents to the spine and neck, and the application of the anode to sensitive points in case these exist. Many nerve sedatives have been used, and also nerve tonics. The exact effect of these seems to be doubtful. In my own case, sedatives, tonics, and electrical applications had all been equally futile to arrest the attacks, but the patient had improved to a considerable degree under the varied treatment. I prescribed galvanism to the spine, arsenic and chloral with some beneficial effect. The hypodermatic use of arsenic under the conditions described by Hammond deserves a trial.

It is useless to discuss the nature of the disease from so few cases as are at our disposal. It has been regarded as a functional neurosis, and to this all must agree, both on account of the absence of any lesion, in one case examined by the most competent neuro-pathologist in Germany, and on account of the absence of symptoms of organic disease and the recovery of the cases. Whether it has a central origin and is produced by a hyper-excitability of the brain or spinal cord, induced by the sudden vaso-motor spasm accompanying fright or mental or physical strain, as Friedreich believed, or whether it may be a reflex spasm due to some peripheral irritation which, being conveyed to the spinal and medullary centres, produces the spasm reflexly, as another author has suggested, remains for the future to decide. The case here reported would seem to favor the latter view.

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(13) Henoch: "Diseases of Children." Wood's Library, p. 81.